396 CT scans from CF patients screened for lung transplant; the Severe Advance Lung Disease (SALD) CT scoring system

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Aim: To design a scoring system to quantify structural abnormalities on the CT scan of CF patients screened for lung transplant with severe advanced lung disease (SALD).
Methods: We collected 57 CT scans from 3 different transplant centers of CF patients screened for lung transplant between 1990 and 2005. First, a panel of 3 experts systematically reviewed a random set of 10 CT scans on eligible items to be used in a first pilot analysis. Included were: bulla/cysts, areas with consolidation/mucous, air trapping, hyper perfusion, and normal lung tissue. For each CT slice and for each of the items the surface area of corresponding lung tissue involved is estimated on a 0–100% scale. Total surface area for the 5 items and each slice adds up to 100%. The final SALD score is a mean volume estimate of abnormal and normal lung tissue.
Results: Pilot analysis of a random subset of CT scans showed a weak correlation between the SALD and Brody score ($p < 0.000$, $R = 0.578$). The intra-observer variability for both scoring systems was good ($Ri = 0.97$). The Brody score was significantly higher in one center compared to the other two ($p = 0.046$ and $p < 0.000$). To our surprise, age had a significant effect on the Brody score with scores decreasing with age ($p = 0.012$). Analysis of the complete set of 57 scans is nearly completed.
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397 Malignant disease adults with cystic fibrosis: a 10 year review

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There are concerns regarding an increased risk of cancer in patients with cystic fibrosis (CF). We aimed to look at the occurrence of malignant disease in a large population of patients with CF.
We reviewed the case-notes of 372 patients attending the Leeds Adult CF Unit. Demographics and data regarding a diagnosis of malignant disease were recorded. A total of 8 patients were diagnosed with a malignant condition over the 10-year period. The median (range) age of diagnosis was 30.5 years (23–55 years).
5 patients were diagnosed with malignancy post-transplantation. Within this group of patients there were 2 cases of lymphoproliferative disease, 1 case of basal cell carcinoma, 1 case of liver cancer and 1 case of small cell lung cancer related to the donor lung. The median time to diagnosis post-transplantation was 2 years (1–6 years) with a median (range) age of 26 years (23–55 years). Within this group 2 deaths were attributed to malignancy.
3 patients were diagnosed with a malignant condition without solid organ transplantation. This group consisted of 1 case of pancreatic carcinoma, 1 case of oesophageal carcinoma and 1 case of bowel carcinoma. The median age of diagnosis in this group of patients was 44 years (27–55 years). Within this group 2 deaths were attributed to malignancy.
Prior to transplantation we found all malignant diagnoses were related to the gastrointestinal tract. Post-transplantation we found a wider variety of malignant diagnoses, with lymphoproliferative disease commonest. Awareness of the increased risk of malignant disease is important and appropriate investigations should be undertaken particularly in patients with atypical symptoms.

398 Renal function in relation to age in cystic fibrosis

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Introduction: The expression of CFTR in the kidney has been demonstrated. In spite of the high expression of CFTR in the kidney, patients with CF do not show major renal dysfunction. Aim: to examine the renal function in CF patients in relation to age.
Patients: 31 stable CF patients (16 male) aged 11 to 34 (mean 19.5±5.2) years were included in the study. CF was diagnosed by two sweat chloride concentrations higher than 60 mEq/l. The patients were divided into Group A: 13 patients, aged <17 yrs, 5 male (mean age 13, range 11–16 yrs) and Group B 18 patients (10 male), aged ⩾17 yrs, (mean age 23.3, range17–34 yrs).
Methods: Markers of glomerular and tubular function were determined: 24-h urine creatinine, electrolytes, albumin and β2 microglobulin, serum urea, creatinine and electrolytes, glomerular filtration rate (GFR) and urinary osmotic pressure (UOP)in a urine sample after water deprivation.
Results: Glomerular Function: GFR was significant higher in group B(94.5–212.6 mean 150 ml/min/1.73 m^2) compared to group A (94.7–141 mean 126.13 ml/min/1.73 m^2) ($p < 0.05$). GFR was greater than 160 ml/min/1.73 m^2 in 50% of group B and none in group A. Tubular function: Microalbuminuria and abnormal level of β2 microglobulin (>40.7 g/mol creatinine), were present in the urine of 4 patients in group B (13%) and none in group A. UOP was low (<800 mOsm) in 7/17 (41.17%) in group B and in 4/13 (30.76%) in group A. All other blood and urine parameters were normal and comparable in the two groups.
Conclusion: Early signs of renal dysfunction were detected in CF adults. Low urinary osmotic pressure was an early indicator of renal dysfunction starting in childhood. Increased GFR may be a sign of hyperfiltration in CF adults. As the life expectancy improves, kidney dysfunction requiring special attention may develop.

399 Evaluation of urinary incontinence in girls and female adolescents with cystic fibrosis

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Introduction: Urinary incontinence (UI) is a known and internationally recognised problem in female adults with cystic fibrosis (CF). UI might have a negative impact on quality of life, but also on lung function, because UI might influence sputum evacuation. The prevalence of UI in young patients (<12 years of age) with CF has not yet been studied extensively. We investigated the prevalence of UI in girls and female adolescents with CF and the influence on quality of life and perceived competence.
Patients and Methods: Questionnaires about micturition symptoms, incontinence, perceived competence and quality of life were sent to all 12–18-year old female patients attending our CF center. Parents of all 6–11-year old girls were only asked to fill out the questionnaire about micturition symptoms.
Results: Fifty-four of 79 patients (68%) completed the questionnaires. The prevalence of UI in the total group was 33% ($n = 18$). Of the 6–11-year old girls 9 of 35 (26%) had UI and 9 of 19 (47%) of the 12–18-year old female patients reported UI. No significant differences in possible risk factors for the development of UI could be demonstrated. Two of 9 adolescent patients reported that UI had an impact on quality of life. We found no differences in perceived competence between the continent and incontinent group.
Conclusion: UI is more common in female CF patients (33%) than would be expected in the general population of females between 6 and 18 years old (2–10%). UI has no impact on perceived competence, but can have impact on quality of life.